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A Study of Contemporary Levels and Temporal Trends in Inbreeding in the Tangier Island, Virginia, Population Using Pedigree Data and Isonymy

RASIKA A. MATHIAS,[1] CAROL A. BICKEL,[2] TERRI H. BEATY,[1] GLORIA M. PETERSEN,[1] JACQUELINE B. HETMANSKI,[1] KUNG-YEE LIANG,[3] AND KATHLEEN C. BARNES[1,2]*
[1]*Department of Epidemiology, The Johns Hopkins School of Hygiene and Public Health, Baltimore, Maryland 21205*
[2]*Division of Clinical Immunology, Department of Medicine, The Johns Hopkins University School of Medicine, Baltimore, Maryland 21205*
[3]*Department of Biostatistics, The Johns Hopkins School of Hygiene and Public Health, Baltimore, Maryland 21205*

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ABSTRACT In this study we describe inbreeding in a large pedigree from Tangier Island, Virginia, in which we compare two commonly used methods to estimate inbreeding in humans: pedigree and isonymy (identical surnames of spouses). Genealogical data on 3,512 individuals dating back to 1722 were used. Using the pedigree method, we determined an average inbreeding coefficient (F) of 0.00873 for the community as a whole, and 0.018 for inbred individuals. Analysis of temporal trends showed that inbreeding began around 1800 and peaked at 0.0109 in 1824–1849 and 1875–1899. Thereafter, inbreeding steadily declined to 0.00565 in 1975–1997. Analysis of pedigree structure complexity over time showed that close consanguinity contributes to inbreeding in the earlier cohorts, and remote consanguinity accounts for much of the inbreeding in the later cohorts. The number of common ancestors increases over time, as does the number of paths connecting inbred individuals to these common ancestors. Inbreeding estimates based on the isonymy approach yielded a 2.2-fold higher value of F (0.01945) compared to the pedigree method. Total isonymy estimates over 25-year cohorts overestimated inbreeding values from pedigree data between 1.5–8-fold. We speculate that the overestimation is probably due to the inability of our data to satisfy the method's assumption of monophyletic origin of each surname. In conclusion, inbreeding in the Tangier Island population is consistent with the isolated nature of its population, and temporal trends reflect patterns in emigration and a breakdown in isolation over time. Am J Phys Anthropol 112:29–38, 2000.

In closed populations with minimal migration, it is not uncommon to observe high levels of consanguinity. Inbreeding is by no means limited to small isolates; low levels of inbreeding have also been observed in large "open" populations (Cavalli-Sforza and Bodmer, 1971; McCullough and O'Rourke, 1986; Czeizel et al., 1976; Jorde and Pitkanen, 1991; Dahlberg, 1938; Sutter and Goux, 1962; Woolf et al., 1956). Scientific interest, however, has been focused on smaller, more

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*Correspondence to: Dr. Kathleen Barnes, 3B65 Johns Hopkins Asthma and Allergy Center, 5501 Hopkins Bayview Circle, Baltimore, MD 21224.

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isolated populations wherein inbreeding has been found to be associated with a pronounced incidence of recessive disorders (Bailit et al., 1966; Hussels, 1969; Ghosh and Majumder, 1979; Pollock et al., 1972; O'Brien et al., 1988; Khoury et al., 1987). Exceptionally high inbreeding coefficients were found by Roberts (1967) in the Tristan da Cunha population (0.0403), Ghosh (1972) in the Kota tribe of India (0.04207), and Bonné (1963) in the Samaritans of Israel (0.043448).

There are four main methods used to measure inbreeding. The most direct method relies upon genealogic or pedigree data. The more indirect methods rely upon isonymy data, migration matrices, or gene frequencies (Jorde, 1989). Several studies have shown that isonymy overestimates inbreeding compared to pedigree data (Jorde, 1989; Rogers, 1987; Hurd, 1983; Roberts and Roberts, 1983; Ellis and Starmer, 1978; Ellis and Friedl, 1976; Friedl and Ellis, 1974; Hussels, 1969). This is primarily due to the fact that most human populations likely violate the underlying assumptions needed for accurate estimation using isonymy. Thus the inbreeding estimated from isonymy is not only composed of a genetic component, but also a component caused due to greater depth in surname ancestry than pedigree ancestry (Lasker, 1991).

Tangier Island is a small island in the Chesapeake Bay comprised of a relatively closed, isolated population. The availability of well-documented genealogical data of the community, dating back to the early 1700s, provides an unusual opportunity to describe inbreeding trends in the context of pedigree structure. Furthermore, the availability of data on the origin of surnames allows us to analyze inbreeding in the context of isonymy, and assess the sources of discrepancy between inbreeding estimates from the two methods.

BACKGROUND OF TANGIER ISLAND

Tangier Island, Virginia, is a 3-by-1-mile-wide marshy piece of land approximately 2 feet above sea level. It lies close to the mouth of the Chesapeake Bay, 12 miles from the nearest mainland port, i.e., Crisfield, Maryland. The island was probably used by Native Americans for hunting and fishing, but was uninhabited. In the late 1600s John Smith claimed all islands in the Chesapeake Bay as part of the Virginia Land Company, naming them the Russell Islands. During the 1700s the Scarborough family, wealthy Maryland aristocrats, held the first land warrant title to the land of Tangier Island. The Scarborough family never lived on the island but rather followed the Colonial landowner custom and grazed livestock on the island to evade the fence post tax, since water surrounding the island negated the need for fencing. Oral tradition names John Crockett as the first settler on Tangier Island in 1686, but this has not been confirmed. The Virginia State records indicate that Joseph Crockett (b. 1722,) son of Sampson Crockett of Maryland, was appointed constable of Tangier Island, and came to live there with his family in 1765.

Contemporary Islanders retain certain Cornish speech attributes reflecting their British heritage, and exemplify the relative isolation of Tangier Island until recent generations; today the island can be reached by a 45-min boat ride or by small aircraft. Limited occupational opportunities have minimized immigration in the 20th century, and contributed to emigration. Episodic events adding to emigration include: 1) an influenza epidemic (1920), 2) hurricane destruction (1933), and 3) military service (World Wars I and II, Korea, and Vietnam).

DATA COLLECTION AND MATERIALS

Genealogical data were collected (1996–1998) using the following sources: United States Federal Census Schedules, 1790–1920, Virginia and Maryland Land, Taxation, Probate, Birth, Marriage, and Mortality Records, Methodist Church birth, marriage, and death records, Church of Latter Day Saints (Mormons) International Genealogic and Locality Indexes, Daughters of the American Revolution (DAR) cemetery records, National Genealogical Society and Virginia State Historical Society resources, Bible records, county histories, gazetteers, personal contacts with Virginia genealogists, and interviews with Tangier Islanders.

TABLE 1. Number of founders for the 10 most common surnames

Surname	Number of founders
Crockett	19
Parks	14
Pruitt	16
Dize	12
Evans	21
Thomas	2
Shores	4
Charnock	2
Marshall	9
Williams	5

Inbreeding on Tangier Island was analyzed using two methods: the pedigree method (Wright, 1922) and the isonymy method (Crow and Mange, 1965; Crow, 1983). The pedigree data were comprised of 3,512 individuals, connected in one large extended pedigree founded in 1722. Individuals were selected by obtaining all known descendants of the earliest founder (Joseph Crockett), and then extending family lines back over generations (a line ends at a founder). The 3,512 individuals comprised 896 nuclear families, ranging in sibships of 1–14.

The data used in the isonymy method were derived from marriage records of the 3,512 individuals. A total of 1,121 marriages was recorded for 2,223 individuals, and 191 surnames were represented. The 10 most frequent surnames are shown in Table 1. A difference in the number of sibships (896) compared to the number of recorded marriages (1,121) was observed. Plausible explanations for this include: (1) a marriage on the island followed by emigration of the couple prior to the birth of a child; (2) marriages without offspring; (3) failure to report infant deaths and still births; and (4) incomplete data collection.

STATISTICAL ANALYSIS

Pedigree method

Inbreeding coefficients for each individual were estimated using the KINSHIP software developed by Boyce. Further analyses of temporal trends in inbreeding were done by partitioning the population into 25-year birth cohorts (1700–1997), resulting in 12 birth cohorts. A total of 106 individuals could not be assigned to any birth cohort because of uncertain birth dates. These subjects were used to assess inbreeding of their descendants, but were excluded from the analysis of temporal trends. The interrelationship between inbreeding trends and pedigree structure was assessed by examining trends in number of common ancestors, number of connecting paths, and average path length for inbred individuals over time.

Isonymy method

In many societies, surnames are transmitted in a manner that reflects biological ancestry. In such cases, surnames may be used to assess inbreeding (Crow and Mange, 1965), and it has been observed that isonymous marriages reflect the same level of inbreeding regardless of the actual degree of consanguinity between the pair (Crow, 1983). For the calculation of inbreeding using isonymy data, both random and nonrandom isonymy should be considered (Jorde and Morgan, 1987).

Random isonymy is that component which estimates inbreeding that occurs simply due to panmixia in a finite population, and can be calculated in two ways:

$$F_r = \left(\sum x_i^2\right)/4, \tag{1}$$

where x_i is the frequency of the i-th surname in the population, including both husbands and wives in the same population; and

$$F_r^* = \left(\sum p_i q_i\right)/4, \tag{2}$$

where p_i is the frequency of the i-th surname in males and q_i is the frequency of the i-th surname in females. Hence F_r^* will equal F_r when the distribution of surnames is not different between sexes (Jorde and Morgan, 1987).

Nonrandom isonymy is the observed isonymy and is also calculated in two ways:

$$F_n = \left(P - \sum x_i^2\right)\Big/\left[4\left(1 - \sum x_i^2\right)\right], \tag{3}$$

where P is the proportion of same name marriages in the population and x_i is defined as in Equation (1); and

$$F_n^* = \left(P - \sum p_i q_i\right) \Big/ \left[4\left(1 - \sum p_i q_i^2\right)\right], \quad (4)$$

where p_i and q_i are defined as in Equation (2) (Jorde and Morgan, 1987).

Total isonymy is obtained by:

$$F = F_n + F_r(1 - F_n) \quad (5)$$

and

$$F^* = F_n^* + F_r^*(1 - F_n^*). \quad (6)$$

Looking at Equations (1–4) it becomes evident that, while the random component of isonymy is always positive, the nonrandom component can be negative (i.e., when P is less than $\Sigma p_i q_i$ or Σx_i^2). The use of isonymy to measure inbreeding has several inherent assumptions, the most important one being the monophyletic origin of surnames (i.e., every name has only one origin) (Crow, 1983). Other problems and assumptions include: 1) common ancestors in the path are assumed to be noninbred; 2) names follow biological inheritance with no allowance for name changes, adoptions, illegitimacy, and errors; 3) nonrandom mating should be symmetrical with respect to sex; and (4) random drift through differential number of offspring must be the same between sexes (Crow, 1983).

In addition to these problems which could give rise to an overestimation of inbreeding when using isonymy, Lasker (1991) also suggests that the overestimation by this method seen in most of the comparative studies also stems from surnames having greater depth in time than the pedigree itself. This "nongenetic" component can be teased apart from the "genetic" component of isonymy using given names, which would perhaps be selected in the same cultural-linguistic fashion as the surnames in the population were originally selected.

RESULTS

Unless otherwise stated, the results presented refer to the 3,406 individuals whose birth dates are known. Figure 1 compares the size of all birth cohorts (inbred plus noninbred individuals) to cohorts of only inbred individuals. Both histograms show similar trends, with a steady increase in population size until 1875–1899. Between 1875–1899 and 1925—1949 the population decreased in size by more than half. Inbreeding did not occur on Tangier until 1800–1824, and prior to 1875, the number of noninbred individuals exceeded that of inbred individuals. The opposite trend was observed after 1875.

Average inbreeding (F) for the whole population is 0.00871 (n = 3,512). The 106 individuals excluded have a minimal impact on the average inbreeding (F = 0.00873, n = 3,406). This is markedly different from the inbreeding coefficient for the inbred individuals (F = 0.01800, n = 1,652). The observed range of F (0.00000–0.15625) indicates the presence of first-cousin matings on Tangier, which were rare and all prior to 1950.

Figure 2 compares the average inbreeding coefficient between 1700–1998 for the entire population and the inbred individuals, respectively. Average inbreeding in the whole population increased from zero before 1800 to 0.0109 in 1825–1849 and 1875–1899. Despite an excess of inbred individuals, inbreeding declined to an average F of 0.00565 in the contemporary cohort. A similar trend was observed among inbred individuals, with an average inbreeding coefficient of 0.0365 in 1800–1824 and 0.0434 in 1825–1849, followed by a steady decline to a low of 0.00973 in the most recent cohort.

To explore temporal trends in the average inbreeding coefficient (F), the percent distribution of individuals by seven categories of F across 50-year birth cohorts was examined (note: 25-year cohorts were collapsed into 50-year cohorts to make trends more prominent). Table 2 demonstrates that in the early cohorts (1700–1849), most individuals belong to the extreme groups (noninbred or highly inbred). After 1850, there is a higher representation in the other categories. However, in every 50-year period, the majority of individuals fall into the noninbred group compared to any of the other six categories. The same trend is observed when looking at only the inbred individuals (data not shown).

These temporal trends indicate that earlier cohorts had a smaller proportion of inbred individuals, but their values of F were high. As the pedigree gained depth over time, the number of inbred individuals exceeds that of noninbred individuals, and

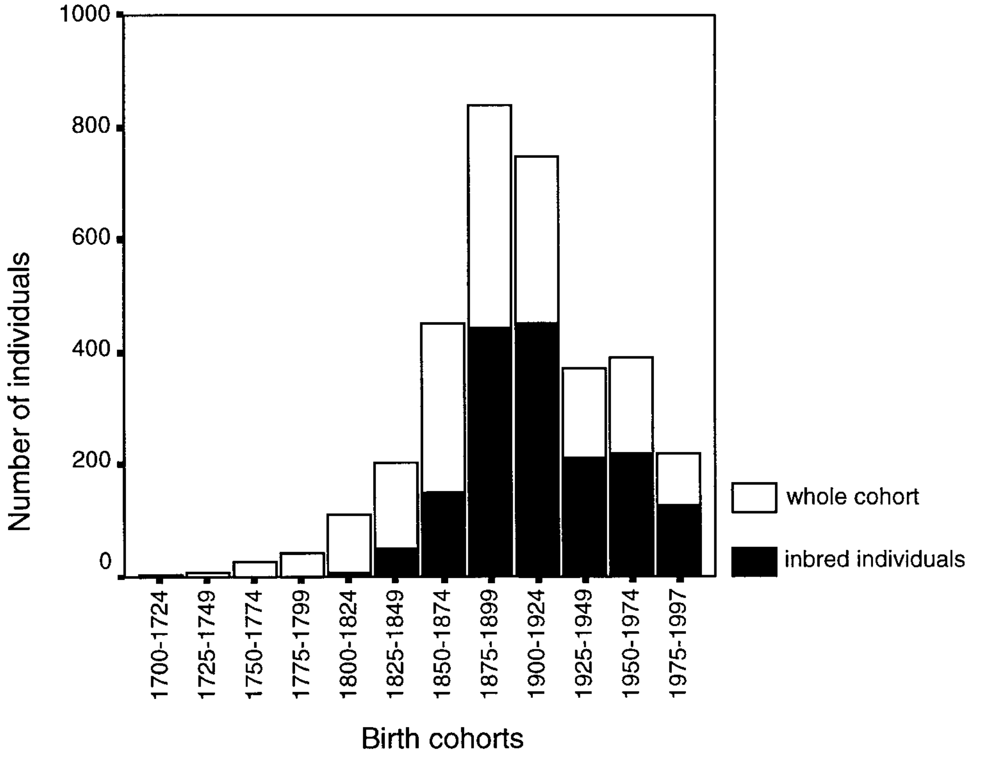

Fig. 1. Histogram of size of the whole population and of inbred individuals over 25-year birth cohorts.

this is accompanied by decreasing values of F. To test the hypothesis that closer consanguinity contributed to inbreeding early in the pedigree, while older remote lines of consanguinity contributed to inbreeding among later generations, we examined three indicators of pedigree structure: 1) number of common ancestors for each inbred individual; 2) number of connecting paths; and 3) average path length. All three indicator variables increase over time, with the most dramatic increase occurring in the number of paths (Fig. 3). It must be noted that the dramatic increase in the number of paths is in part due to the exponential increase in paths (i.e., for every additional ancestor, there are two additional paths).

We examined the average inbreeding coefficient for the inbred individuals over 25-year cohorts in relation to the average contribution of each path to inbreeding (Fig. 4). The average F per path is calculated for each individual (his/her F divided by total number of paths connecting him/her to the common ancestors), and then averaged over the inbred population at that time. After 1849, both factors decrease over time, with a more significant decrease in average contribution of each path to inbreeding. This is because the number of paths increases at a rapid rate over time. These findings suggest closer consanguinity in early birth cohorts, followed by an increase in pedigree complexity and remote consanguinity in later cohorts. Similar findings were shown by O'Brien et al. (1988) in Sottunga, Finland.

Estimates of isonymy were assessed using two methods: 1) treating males and females separately; and 2) combining males and females. Total isonymy for 1,121 marriages on Tangier Island between 1722–1998 is 0.01949 (random component = 0.01954, nonrandom component = −0.00005) treating males and female separately, and 0.01945 (random component = 0.02027, nonrandom component = −0.00083) when

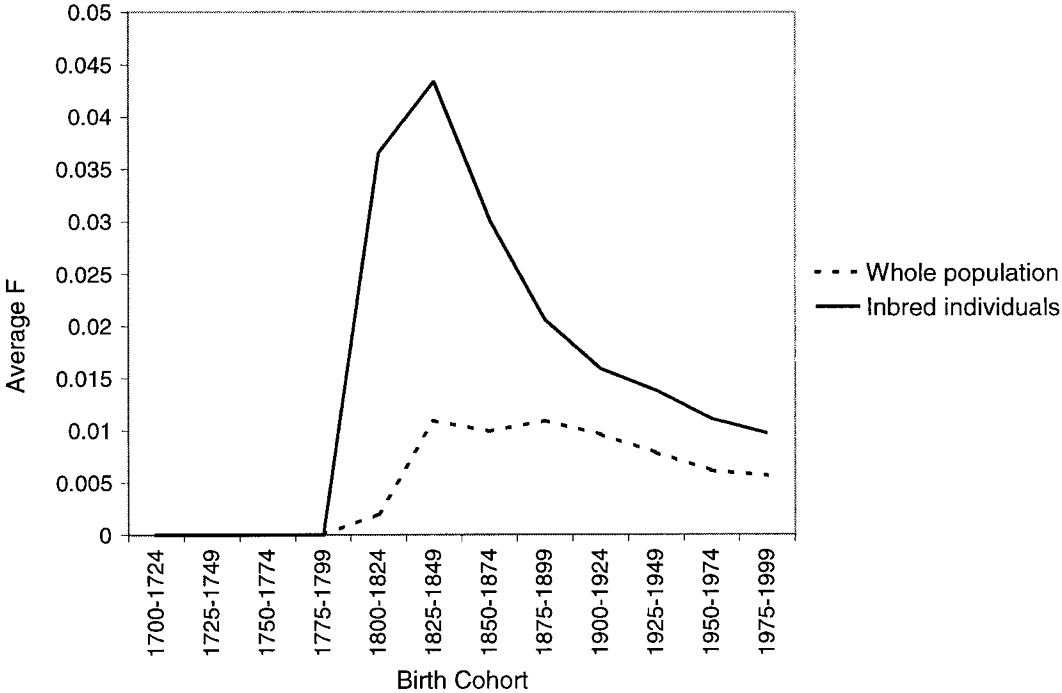

Fig. 2. Average inbreeding coefficient for the whole Tangier Island population and for inbred individuals over 25-year birth cohorts.

TABLE 2. Percent distribution of individuals by categories of F across 50-year birth cohorts

Inbreeding coefficient (F)	Birth cohorts by years						Total
	1700–1749	1750–1799	1800–1849	1850–1899	1900–1949	1950–1997	
0	100	100	81.7	54.3	40.8	43.6	51.5
>0–0.004	0	0	0.0	6.9	10.8	12.2	8.3
0.004–0.008	0	0	0.0	5.3	10.9	10.7	7.5
0.008–0.012	0	0	0.0	6.6	9.7	16.0	8.5
0.012–0.016	0	0	0.0	5.8	3.4	4.9	4.2
0.016–0.020	0	0	0.0	3.3	5.5	6.9	4.3
>0.020	0	0	18.3	17.9	18.8	5.8	15.7

combined. Since there are no differences in the distribution of surnames between the sexes, both estimates are similar. Most of the contribution to isonymy comes from the random component, while the nonrandom tends to be negative.

In this study, average inbreeding for the whole population estimated from the genealogical or pedigree data is 0.00873, while that estimated from isonymy analysis, assuming that males and females have the same distribution of surnames, is 0.01945. The isonymy method overestimates the average inbreeding coefficient by at least 2.2-fold; Figure 5 presents a comparison of the estimates of inbreeding from the two methods over 25-year cohorts (note: the first two cohorts (1700–1724 and 1725–1749) are not included because the number of marriages was small at n = 0 and n = 3, respectively). Isonymy overestimates inbreeding 1.5–8-fold over time. This overestimation is largely due to the violation of the assumption of monophyletic origin of names (Table 1).

DISCUSSION

Tangier Island is an excellent setting to study inbreeding due to its relative isolation and availability of extensive genealogical records. There are no reports of an excep-

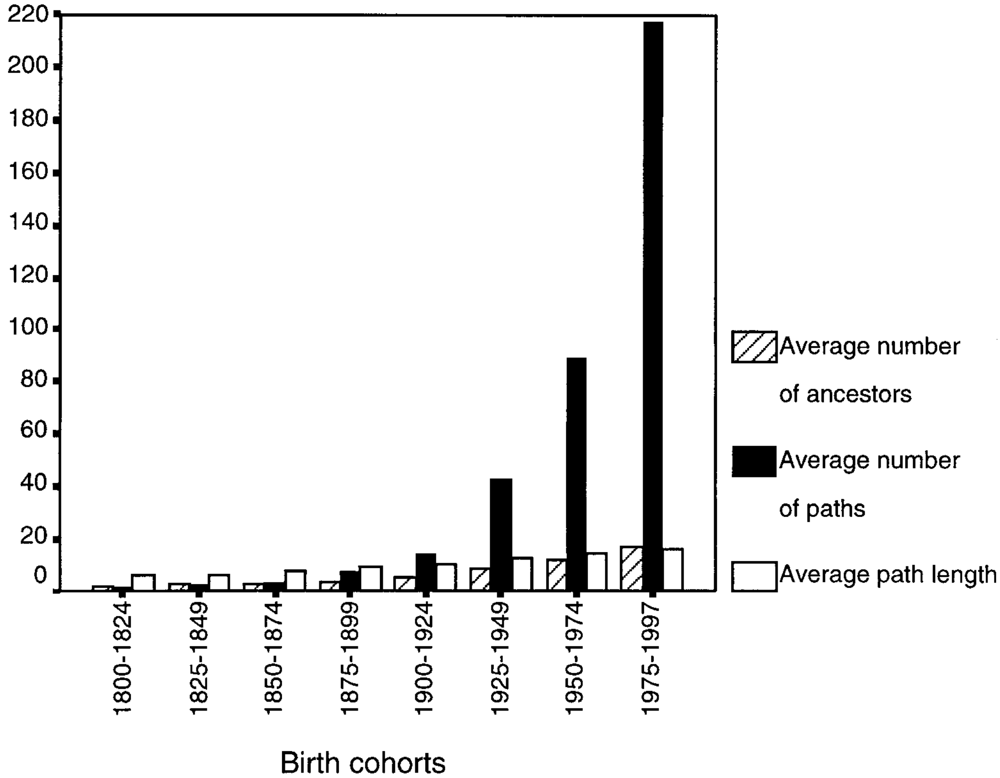

Fig. 3. Trends in number of common ancestors, number of paths, and average path length among inbred individuals in the Tangier Island population over time.

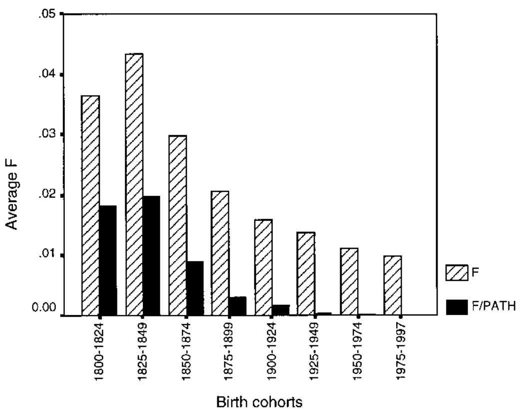

Fig. 4. Trends in average inbreeding coefficient and the average contribution of each path among inbred individuals in the Tangier Island population over time.

tionally high incidence of recessive genetic disorders on the island. Tangier disease, a rare Mendelian disease characterized by low levels of high-density lipoproteins (HDL) in the plasma, was first identified in a boy from the island (Fredrickson et al., 1961). The genetic defect in Tangier disease has been localized to chromosome 9q31 and is believed to be due to a loss-of-function defect (Bodzioch et al., 1999; Rust et al., 1999). However, a greater than expected prevalence of Tangier disease has not been reported on the island since the original case.

The average inbreeding coefficient of 0.00873 on Tangier Island is consistent with genetic isolation and reports of inbreeding in other isolated populations (Bonné, 1963; Hussels, 1969; Ghosh, 1972; O'Brien et al., 1988; Jorde, 1989; de Braekeleer et al., 1992). Increases in emigration are manifested as a decrease in F over time, and are reflected by an average inbreeding coefficient, F, which peaked at 0.0109 between 1825–1849 and 1875–1899, and steadily declined to 0.00565 in the present generation.

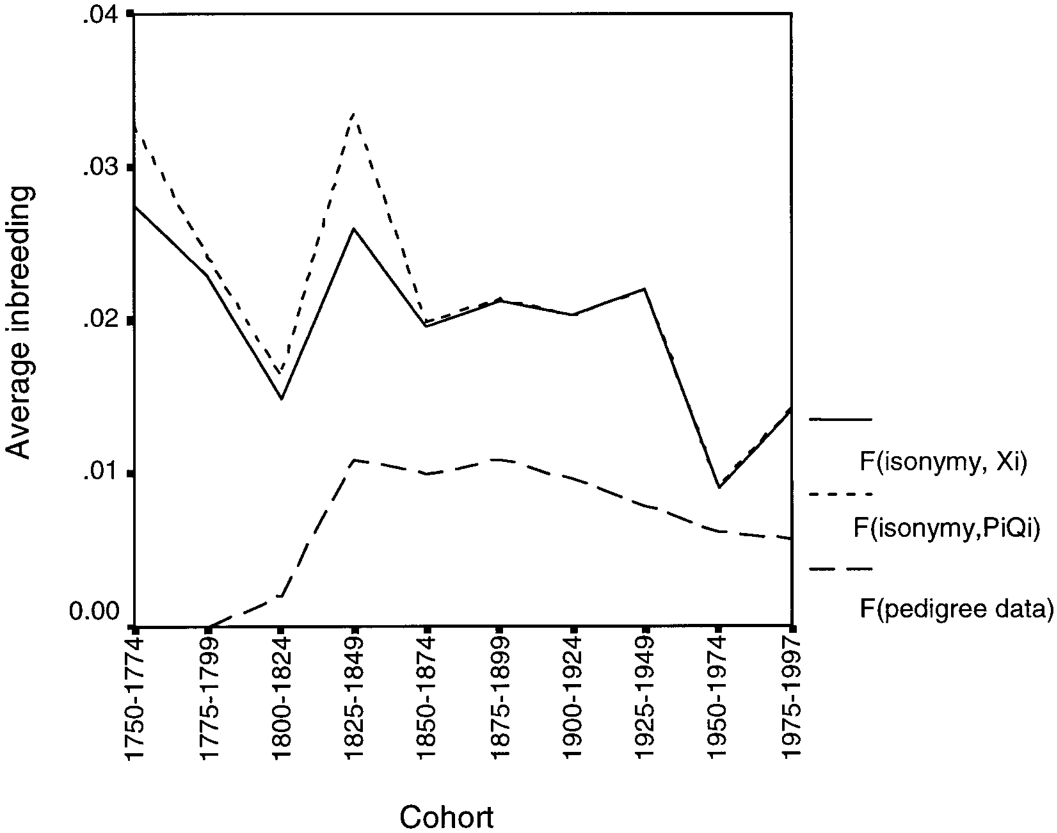

Fig. 5. Plot of estimates of average inbreeding using pedigree data and isonymy methods (X_i, combining males and females; P_iQ_i, males and females separately).

Another explanation for the temporal trends seen in these data is pedigree structure. After the founding cohorts (which are assumed, perhaps erroneously, to be noninbred), earlier cohorts had closer consanguinity with a high average F. In time, the largest contribution to inbreeding came from older remote consanguineous lines of mating. This resulted in declining average F across the birth cohorts, although the proportion of inbred individuals in the population increased. The bias in the average F computed for each cohort, due to data dating back to only 1722, is difficult to assess. Although the direction of the bias is probably toward an underestimation of F, the actual magnitude may be higher in the earlier cohorts, where missing individuals account for a greater proportion of the total population at the time, and where most of the assumed founders are present.

On the island of Tristan da Cunha (Roberts, 1967), the population maintained an isolated state, which is manifested as a steady increase in average inbreeding over time. Roberts (1967) demonstrated that following the founding cohorts, inbreeding on Tristan da Cunha rose, and without any significant immigration, steadily increased over time, with individuals deeper in the pedigree having greater access to common ancestors. In contrast, the Tangier Island population has experienced a steady breakdown in its isolated nature, with an equilibrium between immigration and emigration and mating patterns, that has resulted in a sustenance of low levels of inbreeding in a majority of the population but a decline in average inbreeding.

It is also important to note that the Tangier Island population increased in size until 1899, and within 50 years was reduced to

half its size. This is reflective of the major episodic events of emigration that occurred at this time (an influenza epidemic in 1920, hurricane destruction in 1933, and the World Wars). Following this dramatic reduction in population size and choice of mates, a second sharp peak in average inbreeding would be expected. However, to the contrary, we see a steady decline in the average inbreeding in this population, perhaps indicative of a conscious avoidance of consanguinity in the population. Our preliminary ethnographic observations and unstructured interviews with Tangier residents support the contention that social proscriptions dictating Tangier marriage practices have been observed for at least the past three generations. Informants indicate that a second cousin (preferably "once-removed") is the closest consanguineous relationship acceptable, and the most cited reason for avoidance practices is "because close [relations] have children with medical problems." According to one informant, "[you] grow up knowing who you are related to, and you don't date them."

The observation of an average F of zero until the turn of the 19th century could be due entirely to lack of pedigree depth before this point, since the earliest recorded founder was born in 1722, and his second-generation offspring attained a marriageable age only around 1800. Traditionally on Tangier Island, it is believed that the earliest founder actually dates back to John Crockett in 1686, but the data used in this study start only in 1722. To calculate inbreeding coefficients, we assumed that all founders of the pedigree are not related, which is unlikely in our data. Hence, the average values of inbreeding estimated from our pedigree data will probably underestimate true inbreeding on the island.

This study is consistent with other findings in that the isonymy method overestimates the average inbreeding estimates from pedigree data. In this population, the magnitude of this overestimation was between 1.5–8-fold. Overestimation by the isonymy method is largely due to violation of the assumption of monophyletic origin of names. There are 191 names represented in the data, but 10 surnames accounted for two thirds of all individuals, and each of the surnames had multiple origins. The assumption of monophyletic origin of names is obviously strongly violated among the 66% of the individuals with the 10 most common surnames. Wrongly assuming all of these 10 surnames had only one origin each (i.e., everyone with the same surname is related) clearly leads to an overestimation of inbreeding.

In summary, this investigation has shown that inbreeding on Tangier Island, Virginia is comparable to that in other closed populations, and the temporal patterns of inbreeding on the island reflect increases in emigration and a buildup of pedigree complexity. Further investigation is needed to understand why the reduction in population size between 1899–1950 was not accompanied by any significant changes in inbreeding trends. We are currently comparing observed levels of inbreeding to expected levels to test for avoidance of consanguinity in this population over time. We speculate that social and religious proscriptions in this population strongly influence an avoidance of close mating, and result in an increase in heterozygosity on the island. Perhaps in this time period immediately following the reduction in population size is the strongest avoidance of close mating, and biggest heterozygote excess. The magnitude of inbreeding obtained using the pedigree method is probably an underestimation of actual inbreeding on Tangier Island, while that from isonymy is probably an overestimation. The true estimate of inbreeding on Tangier Island lies somewhere between the estimates from these two methods.

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